

난원공 개존증과 동반된 급성 중간 황반병증

Paracentral Acute Middle Maculopathy with Patent Foramen Ovale

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Purpose: To report a case of paracentral acute middle maculopathy (PAMM) in an otherwise healthy adult with a patent foramen ovale (PFO).

Case summary: A 38-year-old male with no significant medical history presented with acute onset vision loss (20/60) in the right eye. PAMM was diagnosed based on the following findings: hyperreflective band-like lesions at the level of the inner nuclear layer on spectral-domain optical coherence tomography (OCT) and dendritic blacked out areas of hypoperfusion in the intermediate capillary plexus layer on OCT angiography. A systemic workup revealed a large PFO suspicious as a secondary underlying abnormality. After successful closure of the PFO, the pericapillary ischemic signs at the macula were resolved, and the patient's visual symptoms significantly improved to 20/40.

Conclusions: This report verifies the importance of an early cardiac investigation in otherwise healthy individuals with PAMM.

Keywords: Paracentral acute middle maculopathy; Patent foramen ovale; Optical coherence tomography angiography

Introduction

Paracentral acute middle maculopathy (PAMM) is a spectral-domain optical coherence tomography (SD-OCT) finding characterized by band-like hyperreflectivity of the inner nuclear layer (INL) in patients presenting with acute onset visual loss [1]. Localized retinal capillary ischemia at the

parafovea, especially at the level of the deep vascular complex, is a proposed mechanism that is supported by recent findings on OCT angiography (OCT-A) [2]. PAMM may be idiopathic, but it may also be secondary to various retinal vascular diseases and other extrinsic etiologies [3]. Patent foramen ovale (PFO) is a persistent cardiac communication between the left and right atria after birth [4] and can be

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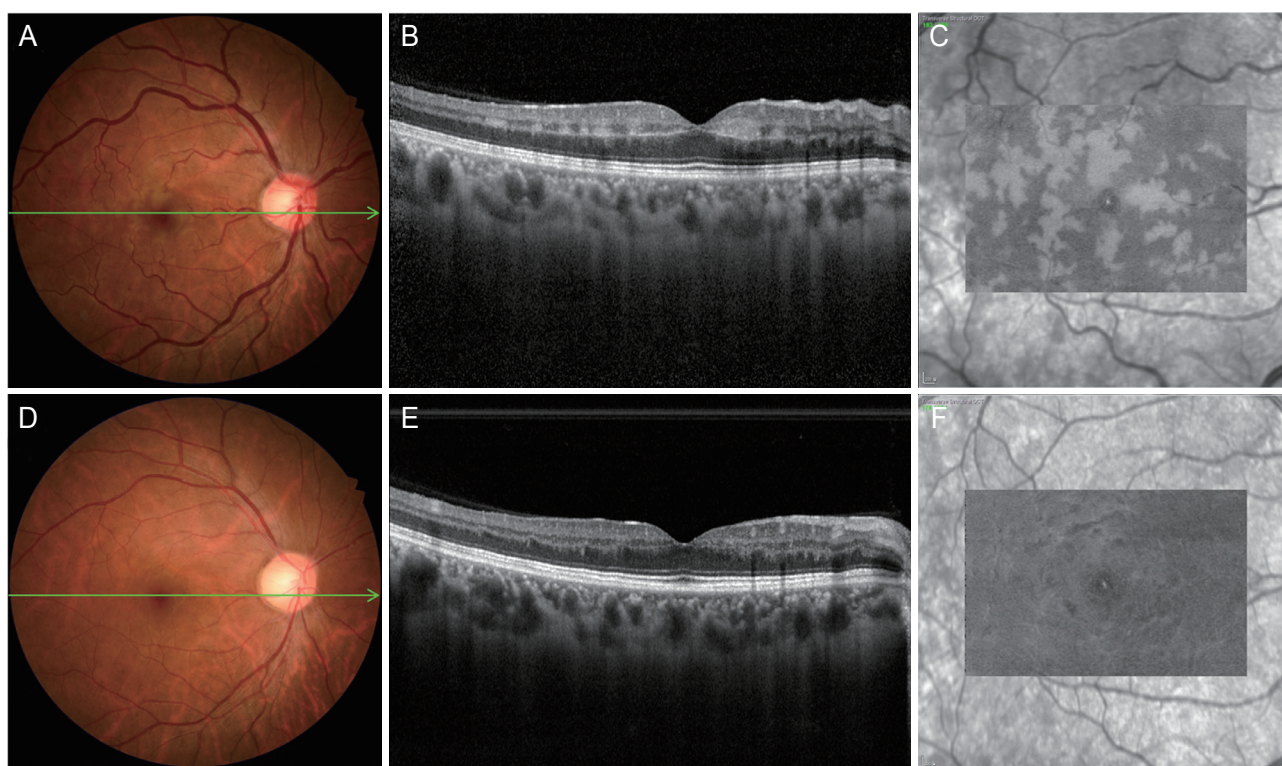


Figure 1. Paracentral acute middle maculopathy in the right eye of a 38-year-old male. Fundus photography demonstrates a fern-like pattern of macular ischemia with dilated tortuous retinal veins (A), and cross-sectional optical coherence tomography (OCT) scans shows multifocal band-like hyperreflective lesions of the inner nuclear layer (INL) (B). Infrared (IR) images (1.9×4.6 mm en face) also reveals hyperreflective dendritic lesions at the baseline evaluation (C). Three months after percutaneous transcatheter closure of the patent foramen ovale, complete regression of macular ischemia signs are noted on fundus examination (D), and the hyperreflective INL lesions disappear, while some remaining localized atrophy is noted at the corresponding sites on OCT (E). The initial hyperreflective lesions on IR imaging had almost completely improve, and hyporeflexive atrophic lesions are faintly observed (F).

associated with rare cases of retinal artery occlusion [5,6]. Herein, we describe a case of PAMM in an adult male patient with a large PFO who presented with acute visual loss.

Case Report

A 38-year-old otherwise healthy male presented with acute onset vision loss in the right eye that started five days previously. Best-corrected visual acuity of the eye was 20/60. His past medical history was unremarkable. Intraocular pressure was in the normal range in both eyes, and the anterior segment appeared normal. Fundus examination showed dilated venous tortuosity and fern-like white retinal patches in the macula (Fig. 1A). SD-OCT showed multifocal band-like hyperreflective lesions of the INL, sparing the outer retina

(Fig. 1B). Infrared (IR) image also revealed highly reflective dendritic lesions at the parafovea (Fig. 1C). OCT-A showed dendritic blacked-out areas of hypoperfusion in the intermediate capillary plexus (ICP) layer (Fig. 2A) which were more prominent in the deep capillary plexus (DCP) layer (Fig. 2B). A systemic workup was performed to investigate the possible causes of ischemia, and all results from the biochemical routine blood tests and coagulation factor tests, including thrombophilia screening, clotting factors, glycoprotein and cardiolipin antibodies, were in the normal range or negative. However, after referral to the cardiology department, the patient was diagnosed with a grade 3 patent foramen ovale (PFO) by a transesophageal echocardiographic bubble contrast study (Fig. 3). The patient underwent successful percutaneous transcatheter closure of the PFO. After three months, his visual acuity increased to 20/40 with concurrent

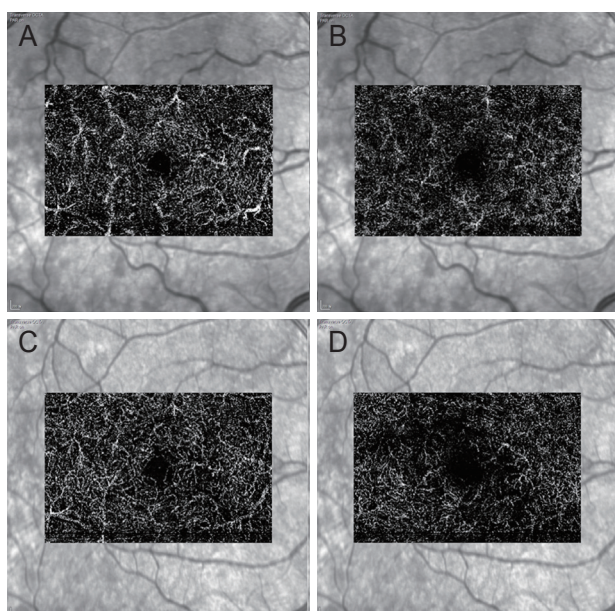


Figure 2. Optical coherence tomographic-angiography (OCT-A) changes in the eye with paracentral acute middle maculopathy. A 1.9×4.6 mm en face OCT-A projection image within the 6×6 mm macular cubes was captured at the level of the intermediate capillary plexus (ICP) (left row) and deep capillary plexus (DCP) (right row). Initial OCT-A images of the ICP (A) and DCP (B) reveal microcapillary hypoperfusion at parafoveal areas corresponding to the lesions noted on the en face imaging. The parafoveal venules are tortuously dilated due to stasis, especially in the ICP. Three months after treatment of the patent foramen ovale, the perfusion defects significantly improve in the ICP (C), but capillary drop outs remained in the DCP (D).

improvement in his visual symptoms. The fern-like white retinal patches had decidedly resolved (Fig. 1D). Moreover, regression of the band-like INL lesions were observed with localized atrophies in the same sites on SD-OCT (Fig. 1E). The hyperreflective dendritic lesions on IR imaging were faintly visible, while the hyporefective atrophic lesions were slightly noticeable (Fig. 1F). OCT-A showed significant improvement of the macular circulation and the perivenular ischemic lesions in the ICP layer (Fig. 2C), while considerable hypoperfusion remained in DCP layer (Fig. 2D).

Discussion

A number of PAMM cases have been reported since its first description in 2013 [1]. The localized grayish lesions at the middle of INL retina, usually in the parafoveal area, is a

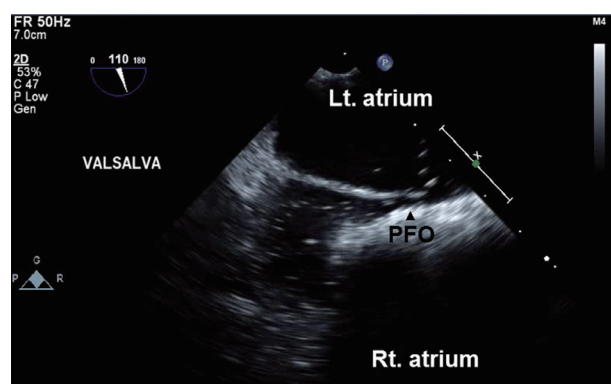


Figure 3. A transesophageal echocardiogram with bubble study shows a right-to-left shunt through a large patent foramen ovale (PFO) (arrowhead).

characteristic finding [3]. Recent OCT-A studies have suggested that deep retinal capillary ischemia is the principal pathomechanism underlying PAMM [7,8].

In the present case, we examined the affected eye by using spectralis OCT-A (Heidelberg Engineering, Heidelberg, Germany) and achieved the adjusted en face images of the capillary networks after manual segmentation of the ICP and DCP layers (superior and inferior to the INL, respectively) according to previous definitions [8]. Drop outs in capillary plexuses and venular stasis appeared slightly more prominent in the DCP than in the ICP. Three months after successful closure of the PFO, capillary hypoperfusion had nearly completely resolved in the ICP but remained in the DCP. This finding may suggest that PAMM involves primarily the DCP, although limitations from OCT-A segmentation may have influenced this outcome and should be taken into consideration.

PAMM may present concurrently with various retinal vascular diseases, such as central retinal vein occlusion, central/branch retinal artery occlusion, diabetic retinopathy, hypertensive retinopathy, Purtscher retinopathy, or retinal vasculitis [3]. Medications, migraines, hypovolemia, orbital compression injury, and/or viral prodromes have also been reported to be associated with PAMM [3]. Therefore, a detailed evaluation of the patient's medical history and a review of the systems are required after the identification of PAMM to investigate the underlying vascular risk factors.

PFO is a persistent communication that allows right-to-left shunting of the cardiac circulation. It is detected in 20-35% of the general population although most patients with an

isolated PFO are asymptomatic [4]. However, the prevalence of PFO can increase the risk of paradoxical embolic events, such as transient ischemic attacks, cryptogenic strokes, and peripheral embolisms. In these cases, PFO closure is considered, especially in younger patients, as a PFO may lead to recurrent thromboembolic events [9]. Central and branch retinal arterial occlusions and optic neuropathy have been described as ophthalmic ischemic manifestations of PFO [6]. PAMM is a sign of microvascular ischemia in the macular area, and thus its association with PFO is possible. Therapeutic options for PFO include percutaneous transcatheter closure, antithrombotic medications, or surgical closure. Transcatheter procedures have proven their effectiveness in the prevention of systemic thromboembolism by safely closing a PFO, like the patient in the present case [10].

To the best of our knowledge, this is the first case report that describes PAMM in the setting of a PFO. If a routine systemic evaluation fails to uncover the underlying cause of PAMM, an exhaustive cardiac evaluation may be helpful, especially in an otherwise healthy adult patient.

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Conflicts of Interest

The authors declare no conflicts of interest relevant to this article.

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